

Case Report

Mixed epithelial and stromal tumor of the kidney: report of a rare case and review of literature

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Abstract: Mixed epithelial and stromal tumor of the kidney (MESTK) is a rare benign tumor composed of epithelial and stromal cells. We report a rare male case with detailed clinicopathological data and follow-up information. The patient presented with gross hematuria. Computed tomography (CT) and magnetic resonance imaging study showed a 60 mm×40 mm cystic lesion with thickened septa and minimal contrast enhancement at the lower pole of the right kidney. The patient underwent nephron sparing surgery (NSS). Intraoperative frozen section showed benign histological features without significant cytologic atypia and mitosis. By additional immunohistochemistry investigations, the epithelial component was positive for cytokeratin-7, high molecular weight cytokeratin, and PAX-8. The stromal component showed strong positivity for vimentin and smooth-muscle actin. This case emphasizes that it is difficult to establish a precise diagnosis of MESTK preoperatively due to lack of any typical radiological features. Thus, intraoperative frozen section is of great clinical significance for NSS with preservation of kidney function. Additionally, regular follow-up is necessary for the MESTK with malignant potential.

Keywords: Kidney, mixed epithelial and stromal tumor, male

Introduction

Mixed epithelial and stromal tumor of the kidney (MESTK) is a rare complex renal neoplasm [1]. It was originally described as cystic hamartoma of the renal pelvis by Pawade et al [2]. The term MESTK was introduced by Michal and Syucek in 1998 [3]. The lesions have strong female predilection, with a 6:1 predominance of women over men [4]. The tumor is typically cystic and solid, which is characterized by a biphasic proliferation of stromal cells with an epithelial component [5, 6]. In this paper, we report the unusual male case of the clinical features of MESTK, and review the related literature.

Case report

Clinical history

A 44-year-old man presented to us with gross hematuria for 16 days, he denied fever, dysuria, and weight loss. Physical examination was unremarkable. He had no significant medical

history of any malignancy and no history of treated with hormone, His routine blood investigations were within the normal ranges. A surveillance magnetic resonance imaging study showed a 60 mm×40 mm cystic lesion arisen from the lower pole of the right kidney (**Figure 1A**). A contrast-enhanced computerized tomographic scan of the abdomen revealed a cystic mass with thickened septa and minimal contrast enhancement (**Figure 1B**). There was no evidence of lymph node or distant metastases. On surgical exploration there was a well-circumscribed tumor mass arising from the lower pole of the right kidney. The patient underwent right nephron sparing surgery. Frozen section revealed a cystic and solid tumor of probably benign nature. In the clinical course, no post-operative complications were observed. The patient was free of local recurrence or distant metastasis until the last follow-up (10 months).

Pathologic findings

On gross examination, the tumor measured 6.5 cm×5.5 cm×5 cm and was observed in the

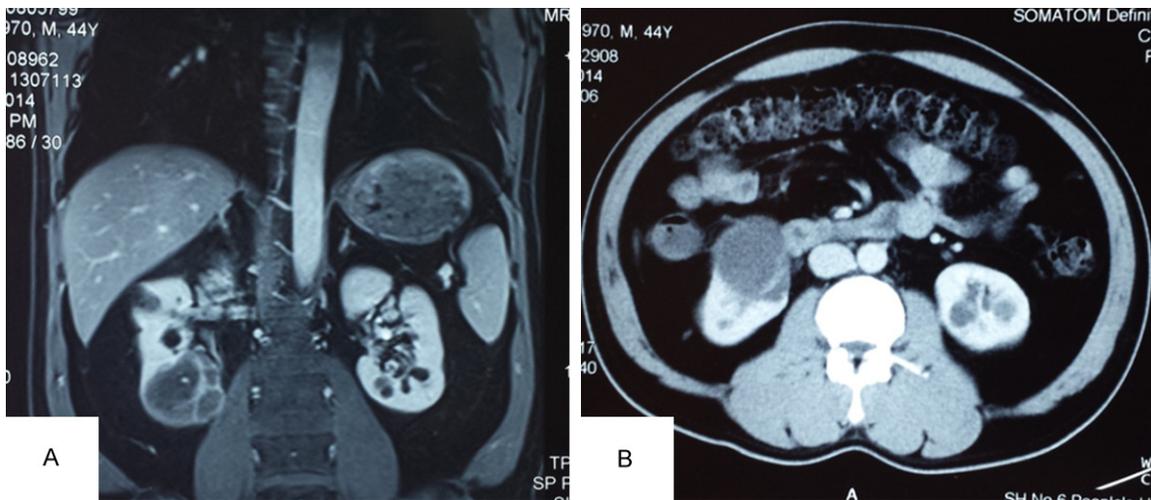


Figure 1. A: Magnetic resonance imaging revealed a cystic lesion arisen from the lower pole of the right kidney. B: Abdominal computed tomography showed a cystic mass with thickened septa and minimal contrast enhancement.

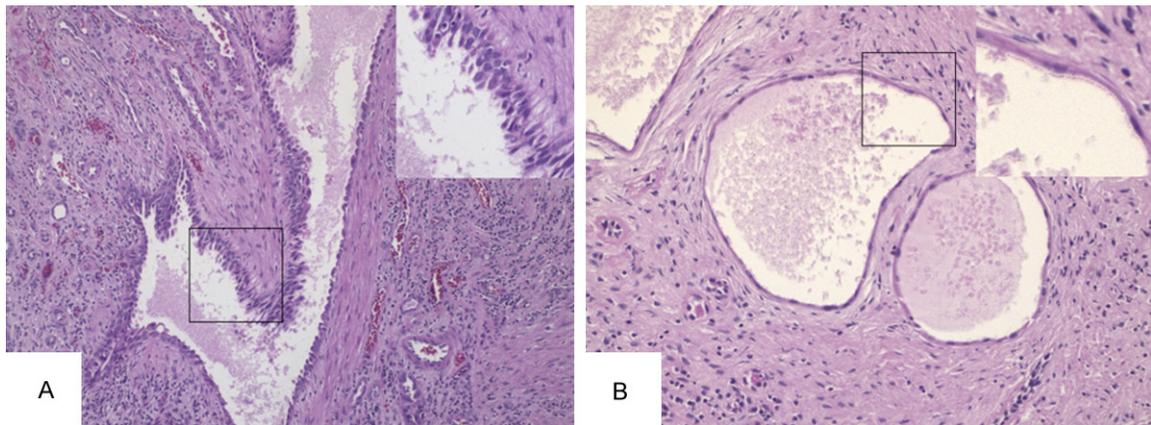


Figure 2. Histologic findings (hematoxylin and eosin, original magnification $\times 100$ [inset, original magnification $\times 200$]). A: The cysts were lined by cuboidal cells with hobnail appearance. B: The lining of the cystic wall was composed of flat epithelium.

lower portion of the right kidney. The mass was well circumscribed and infiltrated into the renal pelvis. The tumor was composed of cystic and solid components with focal hemorrhage and necrosis. Histologically, the lining of the cystic wall was composed of flat or cuboidal epithelium with a focal hobnail appearance (**Figure 2A** and **2B**), and the epithelial cells were relatively uniform and showed no significant cytologic atypia or mitosis. The stroma around the cysts consists of a variably cellular population of spindle cells with no evidence of atypical and high mitotic activity cells. Immunohistochemically, the epithelial lining was positive for cyto-keratin-7 (**Figure 3A**), high molecular weight cytokeratin (**Figure 3B**), and PAX-8 (**Figure 3C**). Spindle cells in the stroma portion showed

strong positivity for vimentin (**Figure 3D**) and smooth-muscle actin (**Figure 3E**), and KI67-positive cells were less than 1% (**Figure 3F**). Both epithelial and stromal components were negative for CD20, CD34, desmin, ER and PR.

Discussion

MESTK is a newly introduced and rare kidney tumor subtype, which was included in the 2004 World Health Organization Classification of Tumors. Antic et al [7] described that MESTK and Cystic nephroma (CN) had similar gross and microscopic features and might represent different morphological variants of the same tumor entity. Hence, Turbinder et al [8] proposed a unifying term of “renal epithelial and stromal

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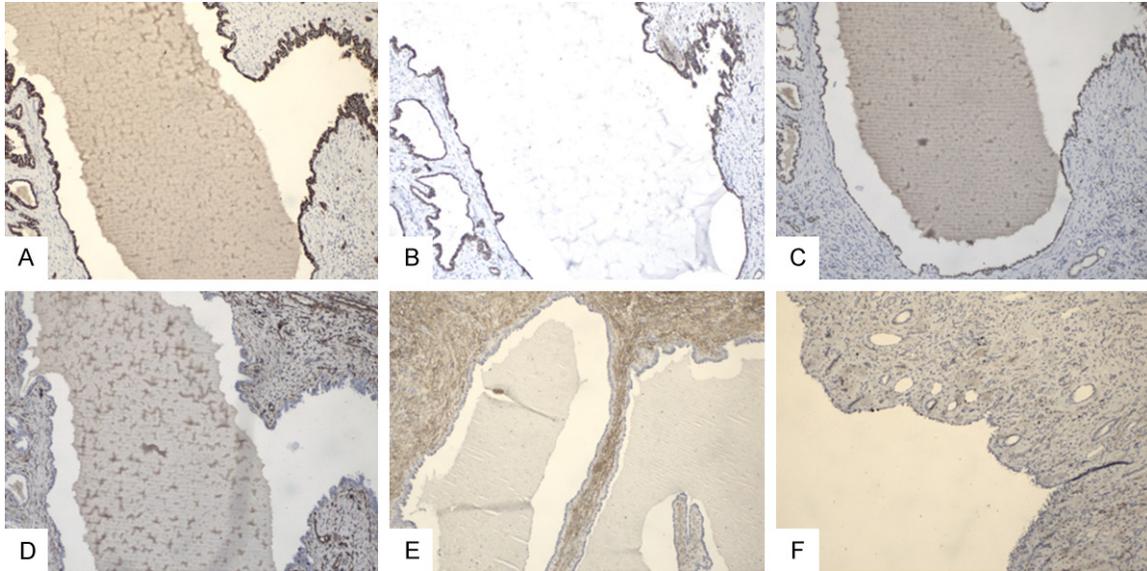


Figure 3. Immunohistochemical staining ($\times 100$) showed that the epithelium was positive for cytokeratin-7 (A), high molecular weight cytokeratin (B), and PAX-8 (C). The stroma demonstrated strongly expression for vimentin (D) and smooth-muscle actin (E), while the stroma was focally positive for KI67 (F).

tumor” (REST) to summarize the spectrum of findings observed in both tumors.

In the current case, the MESTK was composed of both epithelial cells and stromal cells with solid and cystic architecture. The epithelium had features of primitive tubules, and the stroma was variably cellular with areas of muscle differentiation and vascular structures. Radiologically, MESTK appeared as multi-septate cystic mass with solid components and thickened septa on both computerized tomographic scan and magnetic resonance image, and thus could mimic other cystic renal tumors, such as complex renal cyst, cystic nephroma or multilocular cystic renal cell carcinoma [9-11]. It was difficult to establish a precise diagnosis of MESTK preoperatively due to lack of any typical radiological features [12-14], thus, histopathological examination and immunohistochemical studies of the excised renal mass were necessary for postoperatively definitive diagnosis. Immunohistochemical studies revealed the epithelial components were diffusely positive for CK7 and high molecular weight cytokeratin, and the stromal component showed diffusely and strongly positive immunostaining with vimentin and smooth-muscle actin. ER and PR expression used to be described in previous reports of MESTK [4, 15, 16], and histories of estrogen therapy were common in these patients. This implied an underlying associa-

tion between estrogen and MESTK. However, we did not observe any correlation between estrogen therapy and MESTK, as the patient denied a history of hormonal therapy. ER and PR were not expressed in this male patient, which seemed to support the hypothesis that ER and PR expression were not accepted as a supportive diagnostic feature of MESTK and characteristic morphological features should take precedence. Thus, whether the disturbed hormonal environment contributes to the pathogenesis of MESTK warrants further study

Basically, MESTK represents a benign tumor of the kidney, and nearly all cases described so far demonstrated a benign course without tumor recurrence or metastasis. Despite a predictably benign clinical course, a small number of rare aggressive cases with transformation have been recently published in the literature [17, 18]. Our experience suggests that careful intraoperative frozen section examination is required for the choice of operation mode. In order to avoid excessive or unnecessary additional treatment, MESTK must be examined carefully to rule out the possibility of a malignant component.

In summary, the case we presented here showed no recurrence or metastasis until the last follow-up at 10 months. This case emphasizes the fact that the MESTK clinically/radio-

logically mimics other cystic renal neoplasms. Therefore, a pathologic diagnosis of MESTK is of great clinical significance, as MESTK is generally considered to be a benign tumor with good prognosis, and could be cured by surgical excision. In our opinion, nephron sparing surgery (NSS) performed in this case may be appropriate to preserve kidney function. Because the malignant potential of MESTK was recently described, postoperatively continued follow-up might be advisable to observe clinical behavior of MESTK in a subset of these patients.

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Disclosure of conflict of interest

None.

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